MODIFIED FONTAN OPERATION IN FUNCTIONALLY UNIVENTRICULAR HEARTS: PREOPERATIVE RISK FACTORS AND INTERMEDIATE RESULTS

Renate Kaulitz, MD
Gerhard Ziemer, MD
Ingrid Luhmer, MD
Hans-Carlo Kallfelz, MD

Background: The modified Fontan operation has been proposed as definitive palliation for an increasing variety of hearts with complex univentricular anatomy. To eliminate the influence of different surgical strategies, only patients undergoing total cavopulmonary anastomosis were included in this retrospective study. **Methods:** Seventy-two patients had been operated on at ages ranging from 7 to 219 months, with 29 patients younger than 4 years. Twenty-three patients had mean pulmonary artery pressures higher than 15 mm Hg, elevated pulmonary arteriolar resistances ($>3 \text{ U} \cdot \text{m}^2$), or elevated end-diastolic ventricular pressure ($>12 \text{ mm Hg}$). Associated systemic or pulmonary venous anomalies were present in 22 patients (30%), and atrioventricular valve incompetence was present in 21 patients (29%). **Results:** The overall mortality rate was 9.7% (7/72). Variables with significant effects on postoperative mortality were associated systemic or pulmonary venous anomalies, atrioventricular valve incompetence, mean pulmonary artery pressure greater than 15 mm Hg, and prolonged cardiopulmonary bypass time. Postoperative morbidity resulted mainly from atrial arrhythmia (20%). Clinical signs of protein-losing enteropathy or atrial thrombi were rare (3% and 6%, respectively). Postoperative hemodynamic data from 48 surviving patients (74%) revealed a mean transpulmonary gradient of 6.3 mm Hg, systemic venous pressure greater than 12 mm Hg in only 10 patients, and cardiac index less than $3 \text{ L} \cdot \text{min}^{-1} \cdot \text{m}^{-2}$ in 18 patients. **Conclusion:** In a high proportion of hearts with complex univentricular anatomy, associated anomalies, and borderline hemodynamics, the Fontan operation can be performed as a total cavopulmonary anastomosis with acceptable intermediate postoperative morbidity and hemodynamic results. (J Thorac Cardiovasc Surg 1996;112:658-64)

For many years, operations based on the Fontan principle, with separation of the systemic and pulmonary venous return, have been regarded as the definitive palliation for hearts with various forms of functionally univentricular anatomy. Although more complex forms are now being functionally repaired by modified Fontan procedure, the results have

improved significantly, with an overall mortality rate lower than 10%.^{1,2} With increasing experience, many of the preoperative selection criteria defined by Choussat and colleagues³ in 1978 may therefore be exceeded, mainly with respect to associated systemic and pulmonary venous anomalies, atrioventricular valve abnormalities, pulmonary vascular hemodynamics, surgically accessible distortion of the pulmonary arteries, or age at operation.⁴⁻⁶

Most follow-up studies have included a large number of patients who underwent systemic venous-pulmonary artery connection, performed by a variety of techniques including atrioventricular connections and atrioarterial connections (by direct anastomosis or conduit implantation in the pulmonary artery).^{4,7,8} Since 1986, the Fontan procedure with total cavopulmonary anastomosis, which excludes most of the original right atrial wall from the

From the Division of Pediatric Cardiology, Children's Hospital and Division of Thoracic and Cardiovascular Surgery, Surgical Center, Hannover Medical School, Hannover, Germany.

Received for publication Dec. 1, 1995; revisions requested Jan. 26, 1996; revisions received Feb. 23, 1996; accepted for publication Feb. 26, 1996.

Address for reprints: Renate Kaulitz, MD, Division of Pediatric Cardiology, Children's Hospital, K.-Gutschow Str. 8, 30623 Hannover, Germany.

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0022-5223/96 \$5.00 + 0 12/1/73098

Table I. Preoperative risk evaluation

	No.	%
Age at operation <4 yr	29/72	40.2
Systemic or pulmonary venous anomalies	22/72	30.5
Atrioventricular valve malformation	42/72	58.3
Pulmonary artery distortion	7/72	9.7
Mean pulmonary artery pressure >15 mm Hg	10/68	14.7
Pulmonary arteriolar resistance >3 U · m ²	9/68	13.2
Systemic ventricular filling pressure >12 mm Hg	4/68	5.8

Fontan circulation, has been performed in an increasing number of cases.⁹⁻¹¹ Compared with the atriopulmonary connection, this modification was supposed to have the following advantages: reproducibility irrespective of atrial and atrioventricular valve morphology, reduction of the risk of early and late arrhythmias (because less right atrial wall remains at high pressure), low risk of surgical damage of the sinus node artery, and favorable flow patterns within the systemic venous pathway, with reduced risk of atrial thrombosis.^{10, 11}

We report here the early and intermediate results in 72 patients after modified Fontan operation, analyzed with respect to preoperative risk factors and operative variables and their potential influence on intermediate outcome and hemodynamic parameters. To allow a more unbiased evaluation, only patients with total cavopulmonary anastomoses were included in this retrospective study.

Patients and methods

Patients. Since 1988, 72 of 90 patients undergoing Fontan operations for various forms of functionally univentricular hearts underwent total cavopulmonary anastomoses and were included in this retrospective study. Informed consent was obtained from all patients or their parents. Age at operation varied from 7 to 219 months (median 54 months). Tricuspid atresia was the primary cardiac malformation in five patients. Eighteen patients had a double-inlet of left (16) or right (two) ventricle, with or without straddling atrioventricular valves. Forty-nine patients (68%) had other complex forms of univentricular hearts, including left atrioventricular valve stenosis or atresia, common atrioventricular valve, left or right ventricular hypoplasia, and double-outlet ventricle.

Fourteen patients (19%) had such favorable preoperative hemodynamic situations that they had not required any palliative procedures. Most patients had undergone previous shunt procedures. A bipulmonary Glenn anastomosis had been performed after previous shunt operation in 11 patients. Other palliative procedures included those to restrict pulmonary blood flow (pulmonary artery banding in six patients and Damus-Kay-Stansel procedure in

Table II. Preoperative hemodynamic data (n = 68)

	Mean ± SD	Range
Mean PA pressure (mm Hg)	11.9 ± 3.4	5–27
R _p (U · m ²)	1.9 ± 0.7	1–4.2
SV _{ed} (mm Hg)	6.3 ± 2.6	2–15
Qp (L · min ⁻¹ · m ⁻²)	3.7 ± 1.5	1.6–9.4
Qp/Qs ratio	1.0 ± 0.5	0.4–2.9
Arterial oxygen saturation (%)	80.4 ± 6.2	66–97

SD, Standard deviation; PA, pulmonary artery; R_p, pulmonary arteriolar resistance; SV_{ed}, systemic ventricular end-diastolic pressure.

one patient) and resection of subaortic stenosis (two patients).

The decision to proceed with the modified Fontan operation was made on an elective basis in 16 cases of patients who were in good clinical condition and were principally candidates for a Fontan operation. Fifty-six patients needed further operation because of increasingly severe cyanosis or—rarely—congestive heart failure.

Preoperative risk evaluation was based mainly on patients' age, associated anomalies, atrioventricular valve morphology and function, and hemodynamic parameters. Derived from recent publications,^{4, 5, 12} these potential risk factors are summarized for our patient group in Table I. Preoperative hemodynamic variables were derived from cardiac catheterization data (Table II); calculation of pulmonary blood flow and resistance could not be performed in four cases.

Operative variables for modified Fontan operation included additional procedures (resection of subaortic stenosis, procedures for pulmonary artery or atrioventricular valve reconstruction, correction of partial or total anomalous pulmonary venous drainage, additional anastomosis of left persistent superior vena cava to left pulmonary artery), cardiopulmonary bypass time, early postoperative systemic venous atrial pressure, and early and late mortality.

Variables for the intermediate postoperative follow-up included need for reoperation, incidence of late arrhythmias, protein-losing enteropathy and atrial thrombi. Late rhythm disturbances—sinoatrial node dysfunction, supraventricular tachycardia, or atrial flutter—were documented by 12-lead surface electrocardiography or Holter electrocardiography. The New York Heart Association functional class was determined, with some limitations because of the young age of some patients. Postoperative hemodynamic parameters derived from postoperative cardiac catheterization were available for 48 (74%) of the surviving patients at a mean of 28.5 ± 18 months after operation.

Statistical analysis. All data were analyzed with SPSS statistical software system (SPSS Inc., Chicago, Ill.). When appropriate, data were expressed as the mean value (± standard deviation). Numeric data were analyzed with the unpaired Student's *t* test; categorical variables were analyzed with χ^2 analysis. Variables potentially predictive of postoperative death were tested in a stepwise logistic regression model. A *p* value lower than 0.05 was considered significant.

Results

With respect to preoperative risk evaluation, as noted in Table I, 29 patients (40%) were younger than 4 years. Associated systemic (bilateral superior vena cava and azygos continuation in seven and six patients, respectively) or pulmonary venous anomalies (total and partial anomalous pulmonary venous drainage in six and three patients, respectively) were found in 22 patients (30%). Heterotaxia syndrome (asplenia or polysplenia) was identified in seven patients. Atrioventricular valve morphology turned out to be normal in only 30 patients (42%); atrioventricular valve incompetence, as semiquantitatively graded by color-flow mapping, was demonstrated in 29%. Analysis of preoperative hemodynamic data revealed elevated mean pulmonary artery pressure (>15 mm Hg) and elevated pulmonary arteriolar resistance ($>3 \text{ U} \cdot \text{m}^2$) in 10 and nine patients, respectively. The mean ratio of pulmonary blood flow (Qp) to systemic blood flow (Qs) was 1.0 ± 0.5 ; 39 patients had a Qp/Qs ratio lower than 1.0. At least one preoperative morphologic or hemodynamic risk factor was present in 55 of 72 patients (76%).

Surgical results. Associated with the risk potential of early age at operation, nine patients had fenestration of the intraatrial tunnel. Procedures in addition to total cavopulmonary anastomosis were performed in 25 cases (35%), mainly for reconstruction of the pulmonary artery system or additional left superior vena cava–left pulmonary artery anastomosis (nine patients) or resection of subaortic stenosis (five patients). Two patients underwent atrioventricular valve reconstruction or patch closure of the right-sided atrioventricular valve. The early postoperative mean systemic venous pressure did not correlate with either the preoperative mean pulmonary artery pressure or the pulmonary arteriolar resistance.

The overall mortality rate was 9.7% (7/72 patients); five of these patients died during the early postoperative period. All of the patients who died had at least one of the preoperative risk factors determined by regression analysis. Preoperative and postoperative variables were entered into a stepwise logistic regression analysis to assess the combined effects of preoperative morphologic, hemodynamic, and surgical characteristics (Table III). Age at operation, pulmonary arteriolar resistance, and early postoperative systemic venous pressure had no significant effects on overall mortality. Variables with significant effects on

postoperative mortality were associated systemic venous and pulmonary venous anomalies, atrioventricular valve incompetence, mean pulmonary artery pressure greater than 15 mm Hg, and prolonged cardiopulmonary bypass time. Compared with patients without associated venous anomalies, patients with anomalous systemic venous or pulmonary venous connections ($n = 17$) were characterized by older age at operation (94 ± 66 vs 60 ± 48 months, $p = 0.024$), prolonged cardiopulmonary bypass time (221 ± 143 vs 136 ± 36 minutes, $p = 0.028$), and higher early postoperative systemic venous pressure (16 ± 2.8 vs 14 ± 2.8 mm Hg, $p = 0.019$). Analysis of the perioperative data in five of the 17 nonsurvivors (29%) revealed a significant difference from survivors with respect to cardiopulmonary bypass time (382 ± 187 vs 162 ± 47 minutes, $p = 0.04$) and early postoperative systemic venous pressure (19 ± 2.4 vs 15 ± 2.3 mm Hg, $p = 0.015$). When assessing preoperative morphologic and hemodynamic variables, associated pulmonary venous anomalies (as described in 9/72 patients) turned out to have the highest impact on cardiopulmonary bypass time. Surgical results improved significantly with more recent date (calendar year) of operation.

Postoperative morbidity. Sixty-five survivors could be followed up for a mean duration of 48 months (range 12 to 89 months; Table IV). Postoperative arrhythmias, including sinoatrial node dysfunction (seven patients) and supraventricular tachycardia or atrial flutter (six patients), were documented by 24-hour Holter electrocardiography in 20%. There was a trend toward higher prevalence of arrhythmias with increased length of follow-up ($p = 0.048$) but not in correlation with postoperative parameters such as right atrial pressure and cardiac index. Nine patients underwent pacemaker insertion because of sinoatrial node dysfunction or postoperative complete atrioventricular block.

Ten patients underwent reoperation during the first 3 months after operation (including one Fontan takedown); four patients needed pericardiotomy for chronic pericardial effusion and two patients underwent reoperation for residual atrial shunting or anastomosis-stenosis. One late reoperation each was performed for resection of subaortic stenosis and atrioventricular valve reconstruction. None of the patients had clinical or echocardiographic signs of pulmonary venous obstruction.

Two patients had clinical and laboratory signs of protein-losing enteropathy 9 and 38 months after

Table III. Surgical outcome: comparison of surviving and nonsurviving patients

	Total (n = 72)	Survivors (n = 65)	Nonsurvivors (n = 7)	p
Mean age at operation (mo)	68 ± 54	67 ± 52	81 ± 77	0.430
Diagnosis				
TA	5	5	—	0.535
DIV	18	17	1	
Complex SV	49	43	6	
Morphologically SV				
Left	40	37	3	0.476
Right	32	28	4	
Systemic vein return				
Normal	59	56	3	0.004
Abnormal	13	9	4	
Pulmonary vein return				
Normal	63	59	4	0.010
Abnormal	9	6	3	
Atrioventricular valve morphology				
Normal	30	28	2	0.198
Abnormal	42	37	5	
Atrioventricular valve incompetence				
Absent	43	41	2	0.002
Present	21	18	3	
Mean PA pressure (mm Hg)	11.9 ± 3.4	11.6 ± 3.4	14.6 ± 3.8	0.055
>15 mm Hg	10	7	3	0.024
R _p (U · m ²)	1.9 ± 0.77	1.9 ± 0.79	2.1 ± 0.76	0.442
>3 U · m ²	9	8	1	0.918
Q _p (L · min ⁻¹ · m ⁻²)	3.7 ± 1.5	3.7 ± 1.5	3.7 ± 0.67	0.985
Q _p /Q _s ratio	1.0 ± 0.52	1.0 ± 0.54	1.1 ± 0.39	0.550
Arterial oxygen saturation (%)	80 ± 6.1	81 ± 6.1	78 ± 6.9	0.361
Cardiopulmonary bypass time (min)	157 ± 84	139 ± 34	319 ± 189	0.045
Systemic venous atrial pressure (mm Hg)	15 ± 2.4	15 ± 2.3	17 ± 1.7	0.286

TA, Tricuspid atresia; DIV, double-inlet ventricle; SV, systemic ventricle SD, Standard deviation, PA, pulmonary artery; R_p, pulmonary arteriolar resistance.

operation. In addition, one of these patients turned out to have asymptomatic late partial intraatrial thrombosis, as diagnosed by routine postoperative echocardiography. Late postoperative cardiac catheterization in this patient demonstrated poor ventricular function as a result of right coronary artery occlusion; both patients had elevated right atrial pressures of 14 and 16 mm Hg.

Partial atrial thrombosis occurred early and late after operation in three patients and one patient, respectively. Two patients had clinical symptoms of elevated systemic venous pressure or atrial tachyarrhythmia; both patients underwent surgical thrombectomy. The other two patients received thrombolytic therapy, resulting in complete resolution of atrial thrombi.

The general functional status could be described by the New York Heart Association classification in 57 cases; it was sometimes impossible to perform a quantitative exercise test, especially for the large group of young patients. About 65% of the patients for whom functional class could be assessed were in

Table IV. Intermediate results (n = 65)

	No.	%
Arrhythmia		
None	43	66
SND	7	11
SVT	6	9
Pacemaker	9	14
Reoperation	10	16
PLE	2	3
Intraatrial thrombi	4	6
NYHA classification		
I	16/57	28
II	22/57	38

SND, Sinoatrial node dysfunction; SVT, supraventricular tachycardia; PLE, protein-losing enteropathy; NYHA, New York Heart Association.

class I or II. A reliable comparison with preoperative status was not possible.

Forty-eight of the surviving patients (74%) underwent postoperative cardiac catheterization, performed 2 to 75 months after operation (mean 28 months; Table V). With a mean systemic venous atrial pressure of 10.6 mm Hg, none of the patients

Table V. Postoperative hemodynamic data

	Total (n = 48)	Residual right-to-left shunt		p
		Without (n = 38)	With (n = 10)	
Postoperative interval (mo)	28.5 ± 18.7	30.2 ± 18.9	16.5 ± 13.3	0.09
Mean systemic venous atrial pressure (mm Hg)	10.6 ± 2.9	10.9 ± 2.9	9.0 ± 1.0	0.129
Mean pulmonary artery pressure (mm Hg)	10.3 ± 3.1	10.5 ± 2.7	9.3 ± 1.0	0.314
Systemic ventricular end-diastolic pressure (mm Hg)	4.7 ± 2.0	4.8 ± 2.0	4.2 ± 1.7	0.493
Cardiac index (L · min ⁻¹ · m ⁻²)	3.2 ± 0.8	3.1 ± 0.8	3.4 ± 0.5	0.486
Arterial oxygen saturation (%)	93 ± 4.3	94 ± 2.9	86 ± 5.8	0.01

turned out to have a significant gradient at the superior vena cava–pulmonary artery anastomosis; the mean transpulmonary gradient was 6.3 mm Hg. Mean systemic venous saturation and calculated cardiac output revealed normal cardiac indexes (>3 L · min⁻¹ · m⁻²) in 29 patients (60%). Ten patients had systemic venous pressures greater than 12 mm Hg, and 18 had cardiac indexes lower than L · min⁻¹ · m⁻². There were no significant differences in postoperative systemic venous pressures with respect to underlying diagnosis, associated anomalies, and preoperative hemodynamic parameters. There was a trend toward lower postoperative cardiac output among patients with elevated venous pressure. Residual right-to-left shunting (11% to 23%) was related to atrial baffle fenestration (six patients) or to some leakage within the suture lines (four patients), leading to a trend toward higher postoperative cardiac output and lower systemic venous atrial pressure associated with significantly lower arterial oxygen saturation. One patient with significant residual shunting and arterial desaturation of 76% underwent reoperation during the early postoperative period.

Discussion

Preoperative risk evaluation and surgical results. Surgical modifications and increasing experience, especially in the field of postoperative management, allowed a more liberal definition of preoperative anatomic and hemodynamic selection criteria.^{4, 6, 13} In several publications, hemodynamic criteria that indicate increased risk for a Fontan procedure include elevated mean pulmonary artery pressure, pulmonary arteriolar resistance, and systemic ventricular end-diastolic pressure.^{2, 5, 12, 14, 15} Because mean pulmonary artery pressure and pulmonary arteriolar resistance are highly associated with each other and calculation of preoperative pulmonary arteriolar resistance is dependent on pulmonary blood flow, resistance might be estimated as a

second-degree influencing factor. In addition, there is a large variation (from 2 to 4 U · m²) in the upper limit of pulmonary arteriolar resistance considered indicative of higher risk in various studies.* With respect to preoperative hemodynamic parameters, only mean pulmonary artery pressure turned out in our study group to be of significant effect if taken as a categoric variable (>15 mm Hg); increased pulmonary arteriolar resistance (>3 U · m²) and Qp/Qs ratio below 1 were not associated with unfavorable outcome. The number of patients considered in this study is relatively small, however, and it might be possible for additional factors to be assessed as significant in a study with a larger number of patients.

Although Mayer and colleagues⁴ reported no significant effects of anomalies of systemic or pulmonary venous connection on outcome, we accepted these anatomic criteria as preoperative risk factors. Additional systemic venous to pulmonary artery anastomosis (such as in the presence of left persistent superior vena cava) or complex intraatrial baffle techniques necessary to separate pulmonary from systemic venous return in patients with heterotaxia syndrome might lengthen cardiopulmonary bypass time and involve the risk of obstruction of one or both venous pathways. In addition, total anomalous pulmonary venous drainage might also influence preoperative pulmonary vascular hemodynamics. We found a relatively high prevalence of associated pulmonary venous anomalies (in 13% of our patients). Our analysis of potential adverse effects of preoperative variables revealed an especially significant negative effect on mortality of associated systemic and pulmonary venous anomalies. The longer duration of cardiopulmonary bypass in these patients with correction of concomitant anomalies may have compromised postoperative

*References 4, 5, 12, 14, 16, and 17.

systolic and diastolic systemic ventricular function, with consequent increase in postoperative systemic venous pressure. The significantly elevated systemic venous atrial pressure in our nonsurviving patients immediately after operation may be indicative of such an effect.

Although abnormal atrioventricular valve morphology—mainly common atrioventricular valve and left atrioventricular valve hypoplasia—was not a determinant of postoperative mortality in our study, atrioventricular valve incompetence with the need for valve reconstruction had a significant negative effect on surgical outcome. Three of five nonsurviving patients for whom preoperative echocardiograms were available had moderate to severe atrioventricular valve incompetence, which is known to be one of the determinants of postoperative right atrial pressure.

There was no correlation in our study group between age at operation and early or late postoperative mortality. A potential risk of early Fontan procedure as a result of increased pulmonary vascular reactivity was not demonstrable, although 9 patients were younger than 12 months at operation. This may have been an effect of fenestrated total cavopulmonary anastomosis as performed in seven of these patients, which allows maintenance of cardiac output especially during the early postoperative period, when transient myocardial dysfunction or decreased diastolic ventricular function, arrhythmias, or respiratory insufficiency may be more easily overcome.^{12, 18, 19} Because the risk of earlier operation has decreased, most surgeons now recommend the Fontan procedure for 2- to 4-year-old patients,^{4, 5, 13, 20} avoiding additional palliative procedures and preserving ventricular function.

Intermediate outcome and postoperative hemodynamics. With an incidence of 9% to 44%, early and late postoperative arrhythmias are the most important determinants of postoperative morbidity and hospitalization.²¹⁻²⁵ Associated with significant mortality, atrial flutter and junctional ectopic tachycardia turned out to be the most common arrhythmias during the early postoperative period.^{22, 26} In contrast to these studies, none of our patients died because of early postoperative tachyarrhythmias. The overall incidence of late arrhythmias was low, without any significant relation to age at operation, underlying cardiac anatomy, or preoperative and postoperative hemodynamics; however, as described in other studies,²³ early postoperative arrhythmias seemed to predict late arrhythmias. Although late

arrhythmias occurred less frequently among patients after total cavopulmonary anastomosis than among those with atriopulmonary connections,²⁵ patients are still at risk for further decline of sinus node function and development of late tachyarrhythmias.

The rate of early and late reoperations in our study group decreased with more recent date of operation and did not depend on age at operation or underlying cardiac malformation. Because reoperation for relief of subaortic stenosis could pose problems and still carries a significant mortality risk and because increased ventricular mass leads to impairment of ventricular compliance,²⁷ we support myectomy and enlargement of the bulboventricular foramen before or simultaneously with the Fontan procedure.

The protein-losing enteropathy caused by intestinal lymphangiectasia resulting from increased systemic venous pressure usually occurred at least 6 months after the Fontan operation, even in the presence of favorable hemodynamics.²⁸ In our two patients with protein-losing enteropathy, this condition was probably related to long-term elevated systemic venous pressure.

Atrial thrombi are assumed to occur early and late after operation, with a prevalence of 4% to 10%, and in relation to venous stasis and increased systemic venous atrial pressure and size, with potential promotion by coagulation factor abnormalities.²⁹⁻³¹ The relatively low prevalence also reported for our patients may be related to difficulties in diagnosing venous thrombosis by transthoracic echocardiography alone in patients without symptoms; transesophageal echocardiography turned out to have greater sensitivity and is considered mandatory.³² For optimal prophylactic anticoagulation treatment, most centers recommend warfarin for at least 3 months after operation, followed by aspirin.^{33, 34} In all patients with low cardiac output, residual right-to-left shunts, and atrial arrhythmias, however, long-term anticoagulation with warfarin is indicated.^{22, 33}

Even in patients without clinical symptoms, postoperative systemic venous pressure may be elevated to greater than 12 mm Hg at rest. Because systemic venous pressure turned out to be independent of preoperative risk factors, hemodynamic outcome and long-term sequelae of the Fontan circulation are somewhat unpredictable. This unpredictability emphasizes the need for routine postoperative follow-up, including hemodynamic and electrophysiologic aspects as well as detailed laboratory investigations, to discover potential sequelae of the Fontan

operation even in patients clinically free of symptoms. Such a course will allow optimization of long-term medical treatment and consequently of long-term surgical outcome.

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